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Management strategy in patient with familial gigantiform cementoma

A case report and analysis of the literature

Hong-Wei Wang, MD, DDS, Chun-Yue Ma, MD, PhD, DDS, Xing-Jun Qin, MD, PhD, DDS*, Chen-Ping Zhang, MD, PhD, DDS

Abstract

Rationale: Familial gigantiform cementoma (FGC) is a rare benign autosomal dominant fibrocemento-osseous lesion generally limited to the facial bones, typically in the anterior portion of the mandible; it is often associated with abnormalities of the long bones and prepubertal pathologic fractures. Owing to the small number of such patients, a uniform treatment criterion has not been established. This paper presents a patient with FGC who was treated in our department, and offers a systematic review of the patients reported in the literature. Our aim was to explore the treatment strategy for patients with FGC.

Patient concerns: Our patient, a 13-year-old boy, presented with a painless enlargement of the mandible first noted 2 years earlier. It had grown rapidly over the preceding 8 months, affecting both his appearance and ability to chew.

Diagnosis: Based on the pathologic, clinical, and radiographic features, FGC was diagnosed.

Interventions: Mandibuloectomy was performed. The mandibular defect was immediately reconstructed with his right vascularized iliac crest flap. At the same time, a PubMed search was conducted to identify studies reporting on other patients with FGC.

Outcomes: A 3-dimensional computed tomography (3D-CT) scan demonstrated appropriate height of the new alveolar bone. Follow-up results showed recovery of the patient's appearance and mandibular function. He was free of recurrence at 4-year follow-up.

Lessons: FGC is a rare benign fibrocemento-osseous lesion of the jaws that can cause severe facial deformity. Incomplete removal leads to more rapid growth of the residual lesion. Therefore, extensive resection is a suitable strategy to avoid recurrence. Defects of the facial bones found intraoperatively should be repaired with resort to an appropriate donor site. However, it is important to be aware that patients with FGC always have concomitant abnormalities of skeletal metabolism and structure, as well as a vulnerability to fractures of the long bones of the lower extremity. Therefore, the optimal management strategy should include a review of treatment options for other patients as reported in the literature. An optimal protocol can not only provide sufficient high-quality bone suitable for the reconstruction of bone defects, but also minimize complications and maximize quality of life.

Abbreviations: 3D-CT = 3-dimensional computed tomography, CT = computed tomography, F = female, FGC = familial gigantiform cementoma, GC = gigantiform cementoma, M = male.

Keywords: familial gigantiform cementoma, management strategy, mandible reconstruction, vascularized iliac crest free flap

1. Introduction

Familial gigantiform cementoma (FGC) is a rare benign fibrocemento-osseous lesion of the jaw. It is characterized by extensive, well-circumscribed, mixed radiolucent-radiopaque masses in the mandible and the maxilla that can cause severe facial deformity.[1–3] Patients presenting with FGC usually report onset at an early age.[4,5] The goals of treatment are to reset the lesion, preserve the jaw and restore its function, reduce the complications associated with treatment, and improve or maintain the patient's quality of life. However, the management of FGC is difficult owing to its rarity, the rapid expansion of the lesion, widespread involvement of the jaw and high rate of recurrence.[1,2,4,5] Our systematic review of the literature revealed occasional reports of treatment methods for patients with FGC. An important general finding was that incomplete removal of the lesion leads to more rapid growth of the residual lesion. Only extensive resection can avoid a recurrence.[3,4] The repair of osseous defects caused by the surgery is needed as well. We also

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Department of Oral and Maxillofacial-Head and Neck Oncology, Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai Key Laboratory of Stomatology, Shanghai, China.

** Correspondence: Xing-Jun Qin, Department of Oral and Maxillofacial-Head and Neck Oncology, Ninth People's Hospital, Shanghai Jiao Tong University School of Medicine, Shanghai Key Laboratory of Stomatology, No.639, Zhi-Zao-Ju Road, Shanghai 200011, People's Republic of China (e-mail: qinxj1989@sina.cn).*

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noted that, the majority of FGC patients also had lesions in bones other than the jaw.[3,4,6,7] Their histories included prepubertal pathologic fractures of the long bones, which limited the available sources of graft bones for reconstruction of the facial skeleton. On the other hand, the rates of success and complications involving the transplanted vascularized iliac crest flaps were not significantly different from those seen with other microvascular bone transplants.[8–10] For FGC patients, the vascularized iliac crest flap not only provides sufficient high-quality bone suitable for reconstructing segmental mandibular defects, but also minimizes complications. Our patient's treatment is presented in detail in the following paragraphs and the treatment strategy of other reported cases is analyzed. Ethical approval for our work was granted by the Institutional Clinical Research Supervision Committee of our hospital and the informed consent was obtained from our patient.

2. Case report

Our patient, a 13-year-old boy, presented with a painless enlargement of the mandible first noted 2 years earlier. It had grown rapidly over the preceding 8 months, affecting both his appearance and ability to chew. Orthopantography and a computed tomography (CT) scan revealed multiple, expansible lesions mixed radiolucent–radiopaque masses surrounded by a well-defined radiolucent rim throughout the four quadrants of the patient's jaws, typically in the anterior portion of the mandible. The CT scan also revealed polyostotic cortical bone dysplasia. His medical records showed that he had several fractures of the long bones. This patient was diagnosed with FGC based on the pathologic, clinical, and radiographic features and familial history. The treatment program included extensive resection of the lesions and reconstruction of the osseous defect. Considering the abnormalities of long bones and the pathologically tibial fracture (Fig. 1), the vascularized fibular flap was not available. The vascularized iliac crest flap works well for reconstructing defects in the mandible and is associated with an acceptably low rate of morbidity. Therefore, we decided to use a vascularized iliac crest free flap for our patient's mandibular reconstruction.[8–10] SurgiCase CMF software was used to simulate and evaluate the operative procedure. Measurement showed that the defection of mandible up to 10 cm. The preoperative planning software optimized the number and location of bony osteotomies (Fig. 2). Right after mandibulectomy, reconstruction with a vascularized right iliac crest flap was performed (Fig. 3). Follow-up examination with the postoperative 3-dimensional computed tomography (3D-CT) scan revealed appropriate height of the new alveolar bone and acceptable form of reconstructed mandible (Fig. 4). The patient did not accept the secondary insertion of osseointegrated dental implants owing to financial difficulties; however, he was satisfied with the functional and aesthetic results (Fig. 5).

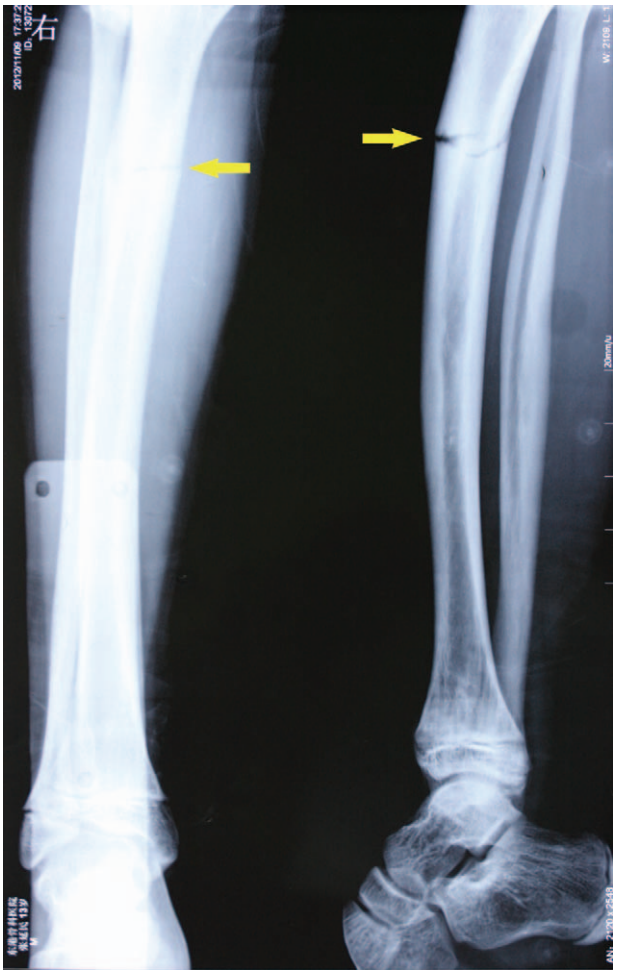

Figure 1. Our patient fractured his left tibia at 13 years of age.

3. Cases analyses

We searched PubMed on November 1, 2016. The search terms were FGC and GC. Upon review of the retrieved articles, cases of GC associated with a positive family history were included. Finally, only 9 articles that fit our inclusion and exclusion criteria were included. These articles resulted in the 55 cases that we included in our analysis (including our case).

The publications retrieved ranged from 1966 to 2016 (PubMed includes articles from 1966 to the present). The largest number of cases published in 1 year were from 1989 (n = 18; 32.73%). The 3 next largest number of cases published in 1 year were from 2015 (n = 15; 27.27%), 1999 (n = 8; 14.55%), and 2008 (n = 6; 10.91%) (Fig. 6). A review of the cases showed that 43.64% (24/55) came from the United States. This was followed by China with 23.64% (13/55). For race, the greatest percentage of patients were Asian with more than 55% (31/55) followed by

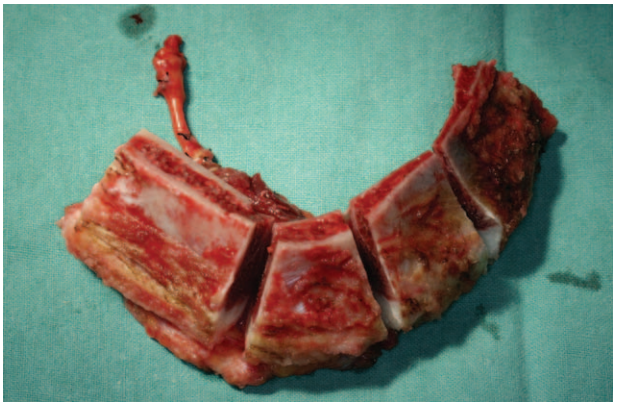

Figure 2. Computer evaluation of the defects and simulation of the reconstruction with 4 sections of iliac bone graft.

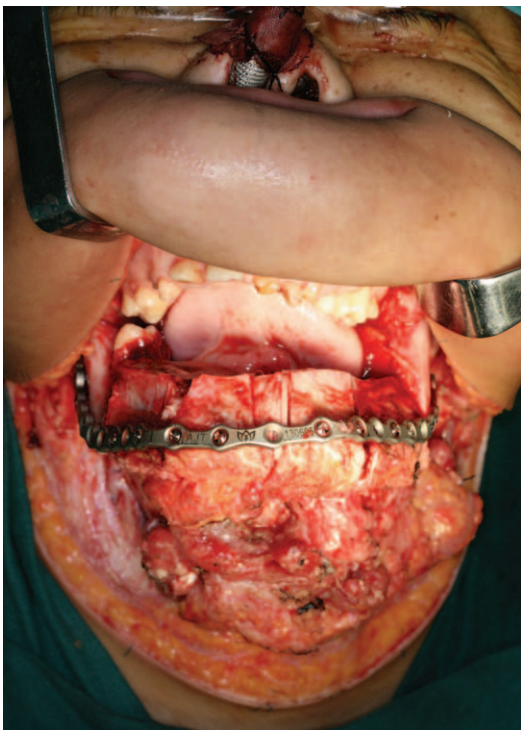

Figure 3. Mandibuloectomy was performed, followed by immediate reconstruction with a vascularized right iliac crest flap.

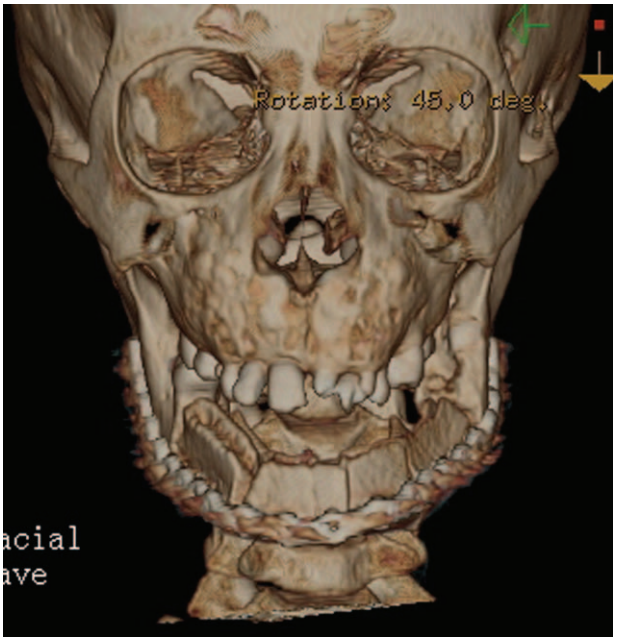

Figure 4. The postoperative 3D-CT scan reveals appropriate height of the new alveolar bone and acceptable form of the reconstructed mandible.

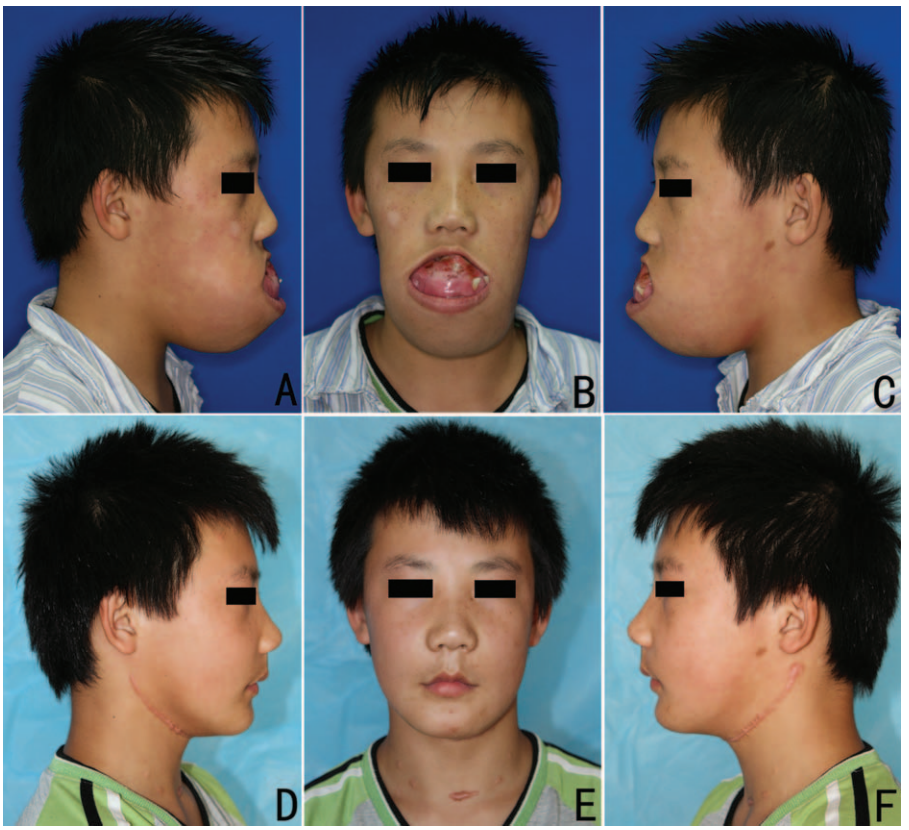

Figure 5. Preoperative frontal and lateral profile photographs (A–C) and postoperative frontal and lateral profile photographs (D–F). These images (D–F) show satisfactory aesthetic results.

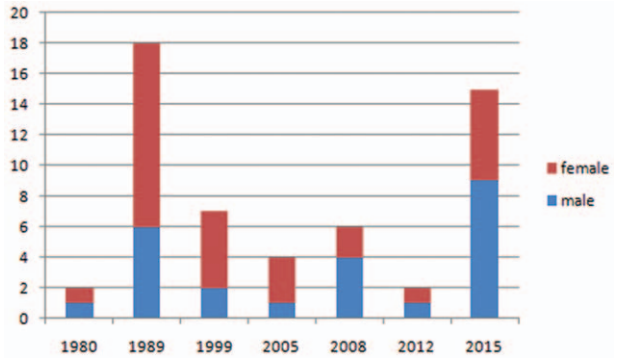

Figure 6. Number and gender of cases according to year of publication.

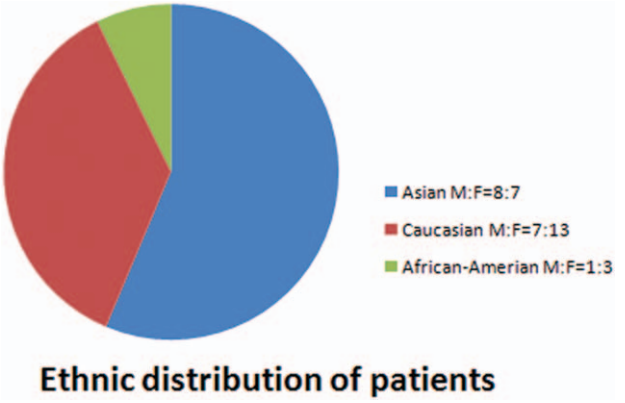

Figure 7. Ethnic distribution of patients and sex ratio in each race.

Caucasians with almost 40% (20/55). The proportion of African-American was less than 8% (4/55) (Fig. 7). In this series, 24 patients (43.64%) were male and 31 (56.36%) were female. Information on treatment was gathered from 21 cases, of which 19 (90.48%) underwent surgical resection. Facial bone reconstruction was performed in 5 patients. Table 1 shows the publication date, nationality, race, and treatment variables of these cases.

Table 1

The nationality, race, sex, and treatment of patients, as well as the article publication dates of reports.

Refs.	Nationality and race	Number of cases	Sex	Number of treated patients	Detailed information on treatment
Cannon et al[12]	America and Caucasian	2	M:1	2	M: Partial excision with aesthetic recontouring was done in 3 separate procedures
			F:1		F: Twelve surgical procedures in the mandible and maxilla were performed in an attempt to control the original and recurrent lesions
Young et al[1]	America and Caucasian	18	M:6	6	Five patients have had partial mandibulectomies
			F:12		One patient has had cosmetic shaving of the maxilla, which only in regrowth of the tumor at an accelerated rate
Finical et al[2]	Philippines and Asian	8	M:2	5	Extensive resection of the tumors and complex reconstruction of both the soft tissues and facial skeleton. The average number of operations required to treat these patients was 7
			F:6		F: This patient's entire mandible was resected and replaced with a reconstruction plate. Four years later, she resected the remaining tumor involving the left maxilla, nose, and left orbit was resected and the area reconstructed with rib grafts. At the same time, the patient's free-floating mandibular reconstruction plate was removed and a total reconstruction of the mandible using bilateral fibular free tissue transfers was performed
					F: This patient's tumor was resected and the maxilla reconstructed with autologous rib bone grafts and the mandible was replaced with a large reconstruction plate
					M: This patient's tumor was resected and the maxilla and nose were reconstructed with autologous rib bone grafts
Rossbach et al[3]	America and African-American	4	M:1	2	F: Resection of a slowly growing mandibular mass
			F:3		F: Partial mandibulectomy
Moshref et al[6]	Iran and Asian	6	M:4	2	M: A partial maxillectomy was performed, and oral rehabilitation was accomplished with a partial removable denture
			F:2		M: Alveoloplasty with curettage of the necrotic bone was carried out and the patient was referred for prosthodontic rehabilitation
Shah et al[5]	Korea and Asian	2	M:1	2	M: This patient underwent 3 surgeries in his second decade
			F:1		F: Five surgical excisions of the original and recurrent lesions on both maxilla and mandible were performed, from 2.5 to 6.5 years of age
Wang et al[4]	Chania and Asian	13	M:8	2	M: This patient underwent segmental mandibulectomy. The mandible was reconstructed with a free iliac crest flap
			F:5		F: This patient underwent segmental mandibulectomy. The mandible was reconstructed with autologous nonvascularized iliac bone grafts. Thirty years later, the patient underwent subtotal maxillectomy and the maxillary structure and function were restored with a removable prosthesis
Şakar et al[11]	Turkey and Asian	2	M:1	2	M: Owing to a surgical contraindication, a tooth-supported maxillary complete denture to provide lip support and a mandibular overlay removable partial denture (ORPD) were selected as the alternative treatment
			F:1		F: Tooth-supported complete dentures were used to provide lip support and rehabilitate the decreased occlusal vertical dimension

F = female, M = male.

4. Discussion

FGC, a rare familial form of cementoma, is considered a benign fibro-osseous lesion; it follows an autosomal dominant inheritance pattern with variable phenotypic expression.[1–6] It was first reported in 1953 by Agazzi and Belloni, who described it in an Italian family. Young et al in 1989[1] suggested that it should be recognized as a separate entity. The literature is limited to isolated case reports and small case series. Patients associated with a positive family history are extremely rare. After searching PubMed, we found that since the first publication in 1953, there appears to have been a total of 55 cases, which most commonly appeared in Asia.[1–7,11,12] This was followed by North America. FGC typically presents in the first or second decade of life, with a predilection for Asian and Caucasian ethnicities. Among these cases, 24 were male and 31 female. There was a slight female preponderance. We also noted the ethnicity-related gender ratio difference, with a slight male preponderance in Asian and an obvious female preponderance in Caucasian and African-American patients. Because of the low number of patients, further statistical analysis is required in the future.

FGC is generally limited to the facial bones, typically in the anterior portion of the mandible.[4,5] This frequently manifests with poorly fitting dentures, interdental spacing, and difficult mastication. Our literature review demonstrates that 19 patients underwent surgical resection, in most instances a partial excision.[1–7,11,12] But simple recontouring of the lesions or incomplete removal accelerates the growth of the residual lesion and leads to recurrence.[1,4,5,12] In the case of a 6-year-old child, cosmetic shaving of the maxilla resulted only in regrowth of the lesion at an accelerated rate.[1] In a 20-year-old man, partial excision with recontouring was performed in the area of the mandibular symphysis. Two years after surgery, Cannon et al[12] found that the involved areas of the maxilla and posterior portion of the mandible in this patient, which had not been surgically manipulated, had clinically and radiographically remained unchanged, but significant symmetric regrowth had occurred in the area of the mandibular symphysis. Therefore incomplete removal or simple recontouring of the lesions for aesthetic reasons or to facilitate the wearing of a dental prosthesis is not recommended.

To achieve radical cure and avoid recurrence, complete surgical excision is needed in patients with FGC.[1,2,4–6] In a 48-year-old Iranian male, for example, a partial maxillectomy was performed and oral rehabilitation was accomplished with a partial removable denture.[6] In cases involving significant bone defects and pronounced facial asymmetry, facial reconstruction is needed following extensive resection of the lesion.[2,4]

A variety of options have been successfully used for facial bone reconstruction. The autogenous nonvascularized fibula, rib, and iliac bony flap has been important resource for the reconstruction of jaw defects, however it is better to use these flaps to reconstruct small bony defects that will not be irradiated after surgery.[13,14] They cannot offer enough bone for complex reconstructions.[13] The later absorption of the grafted bone also have seriously affected the patients' appearance and function. To date, facial bone reconstruction was performed in 4 FGC patients (exclude our patient).[2,7] Three of them had their maxillary lesions resected. The defects of maxilla, nose, and orbit were reconstructed with autologous rib bone grafts. The postoperative appearance at 9 months was satisfactory.[2] One patient underwent subtotal maxillectomy; her maxillary structure and function were restored with a removable prosthesis.[4] In 3 patients, a reconstruction plate, fibular free tissue, and autogenous nonvascularized iliac bone were used for mandibular reconstruction. One patient was a 37-year-old woman, whose entire mandible was resected and replaced with a reconstruction plate. Four years later, the free-floating mandibular reconstruction plate was removed and a total mandibular reconstruction with bilateral fibular free tissue was performed.[2] Another woman, our patient's aunt, underwent resection of the mandibular mass and reconstruction using an autogenous nonvascularized iliac bone graft. However, her 30-year postoperative 3D-CT scan showed obvious absorption of the iliac bone, which had seriously affected the patient's appearance and ability to chew. Therefore, the autogenous nonvascularized bone cannot be used for the reconstruction of large mandibular defects in patients with FGC.

With advances in microsurgical techniques, free vascularized bone grafts, such as the scapular flap, fibular flap, and iliac crest flap, have become the preferred grafts for the reconstruction of jaw defects.[10,15–18] Mandibular reconstruction with a scapular flap has not been popular because this flap does not supply a sufficient amount of bone.[15–17,18] In recent years, use of a vascularized fibular osteocutaneous flap has become standard for reconstruction of the mandible.[19] Fibular flap has a long length of good-quality bone and a sufficient blood supply suitable for stable osteosynthesis. The perforators of the peroneal artery permit the inclusion of a reliable skin paddle in the fibular flap for better reconstruction of the soft tissue.[17,20] However, one weakness of the fibular flap is that the height of the reconstructed mandible is lower than that of the iliac crest. The dimensions and shape of the iliac crest are similar to those of the lateral mandible. It can offer a large amount of bone for complex reconstructions in the dentate mandible.[9,10,21] Osseous or osteocutaneous flaps can be harvested depending on the volume of the soft tissue defect.[16,18] The rates of success and complications associated with the use of vascularized iliac crest flap transplants are not significantly different from those of other microvascular bone transplants.[8–10] It even has a higher rate of osseointegration, thus enabling implant placements for later prosthetic treatment.[10,18] This implants therefore facilitate good reconstruction and further improve the patients' quality of life. For patients with FGC, long bone dysplasia, and prepubertal pathologic fractures, especially tibial fractures, limit the use of fibular flaps. Therefore, the vascularized iliac crest flap is the best choice for mandibular reconstruction. Because of our patient's multiple femoral fracture and tibial fractures, we chose to use a vascularized iliac crest free flap for his mandibular reconstruction. The postoperative 3D-CT scan revealed an appropriate height of the new alveolar bone and an acceptable form of the reconstructed mandible.[4]

For some special patients, extensive resection is not recommended owing to surgical treatment contraindications. In order to restore their oral function and maintain their quality of life, removable partial denture can be selected as an alternative treatment. Şakar et al[11] reported on 2 patients presenting FGC and Ehlers–Danlos syndrome. Tooth-supported complete dentures and overlay removable partial denture were used for their treatment. No muscle tenderness, tooth sensitivity, or temporomandibular dysfunction was observed during 1 year.[11]

5. Conclusions

For FGC patients without surgical contraindications, complete surgical excision is the most effective strategy for treatment. Oral rehabilitation and facial reconstruction are needed following extensive resection of the lesions. Because of concomitant long

bone fractures, that compromise their use, the iliac crest is a well-established donor site with adequate tissues and an acceptably low rate of complications. The vascularized iliac crest flap can be contoured by osteotomies and so as to allow anchorage of dental implants to complete the patient's reconstruction and rehabilitation.

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